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CONGENITAL DEXTROCARDIA
COR TRILOCULARE BIVENTRICULARE
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M.B., C.M., 1893.



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The aim of the present paper is to describe a case of congenital heart disease which showed the above characters and in which a confirmatory post-mortem was obtained. Thereafter it is proposed to discuss the details of the case and pass in review such matters as seem to arise from these individual characters as well as their bearing on each other and on their etiology.

The patient in question died at the age of $2 \frac{10}{12}$. His life history until two months previous to his death was as follows -

He is a boy of $2 \frac{8}{12}$ ths. who has suffered from this cardiac anomaly since birth.

His parents are healthy, and show no evidence of rheumatism, heart disease or syphilis. They have one other child, a healthy girl of five. The great grandfather, the grandfather and two grand aunts all on the maternal side suffered from heart disease. The father's brother had rheumatic fever at the age of seven and was subsequently ill with heart disease until the age of fifteen when he died of heart failure. There is thus very complete evidence of the tendency to rheumatic /

rheumatic and cardiac affections on both sides although as yet the child's parents have escaped. The child was born at full time in the normal way but it was noted from the first that he was suffering from some cardiac affection. For the first day or two he was semi-conscious, did not cry much and looked as if he would not survive long. Gradually he came to himself although even then signs of dyspnoea showed themselves. The first year of his life was one long struggle with existence. Dyspeptic and bronchitic troubles were added from time to time to his heart affection. After much experimenting a suitable dietary was hit upon and thereafter a slow but yet decided improvement set in although not a few relapses occurred. Fainting fits were frequent in the earlier months - generally of short duration but at times lasting for half an hour. These 'turns' varied greatly in character from a slight unconsciousness with cyanosis to a more evident fit with general stiffness of the limbs. In time these ceased almost altogether but with teething they were again called forth. His second year was much freer from such incidents. He now began to grow rapidly in length though there was very little gain in weight. Speech came early but he did not walk till 2 3/12ths. The /

The cardiac condition has remained unchanged.

At the age of 2 8/12ths he is now a poorly developed child and distinctly under weight 26 lbs. There is slight rickets with beading of the ribs and transverse constriction of the chest. He takes His food well, runs about a little and seems fairly comfortable. At the same time he has an anxious face with slightly distended veins on it. He is quite active and bright. There is a little cyanosis about the lips. The veins in the neck are full and prominent. There is slight clubbing and congestion of the fingers and toes. His mother has noted that his fingers and toe nails and also the hair of his head show a more rapid growth than usual. There is no oedema anywhere. The veins over the front part of the chest are distended.

The pulse is 100, small and regular. There is evident pulsation over the right side of the chest. It is chiefly marked in the 5th and 6th interspaces just below the right nipple. To palpation it feels strong, regular and forcible. There is no accompanying thrill. The pulsation is widely felt over the chest. There is pulsation in the vessels of the neck just following this pulsation of the presumable apex beat /

beat. The epigastrium shows a pulsation synchronous with it. There is no pulsation over the left side of the chest.

On percussion the limits of the heart are seen to be 1 upper border at 2nd rib on the right side:

2 right border midway between right arteria axillary line and right nipple line and 3 left border at the right sternal edge. The lower border blends with the underlying liver see diagram On auscultation the main feature is a loud rough systolic murmur replacing the first sound. This murmur is heard all over the heart area and outwards into the right axilla. Its maximum point is at the apex beat. The second sound is loud and accentuated but of late it seems to be replaced by a diastolic murmur.

An x-ray photograph of the chest shows the right side of the chest with a dark shadow corresponding to the heart on this side. This dullness extends upwards to the level made out by percussion and below blends with the liver. There is no change in the level of the diaphragm on the two sides. see x-ray photograph Over the usual heart area there is absence of apex-beat and the note is resonant while on auscultation there is merely heard the conducted weakened sound from the right side. /

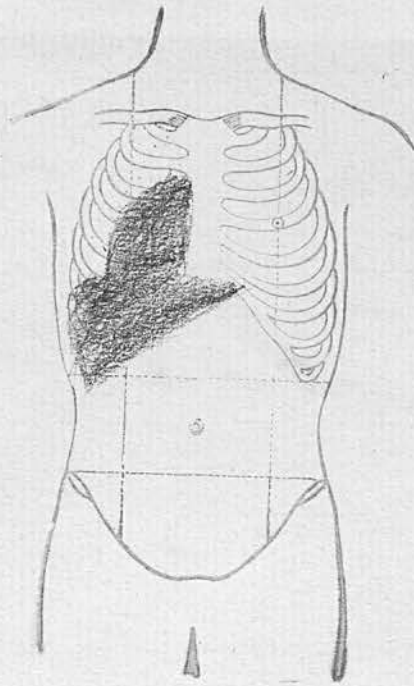


Diagram showing the position and size of the heart and liver as determined by percussion.



Left

Right.

View from back.

X-Ray Photograph of Child (taken when about 2 years old). There is some little movement but there is quite evident dullness over the right side with the axis of the heart directed downwards and to the right. The diaphragm shows much the same level on the two sides.

side.

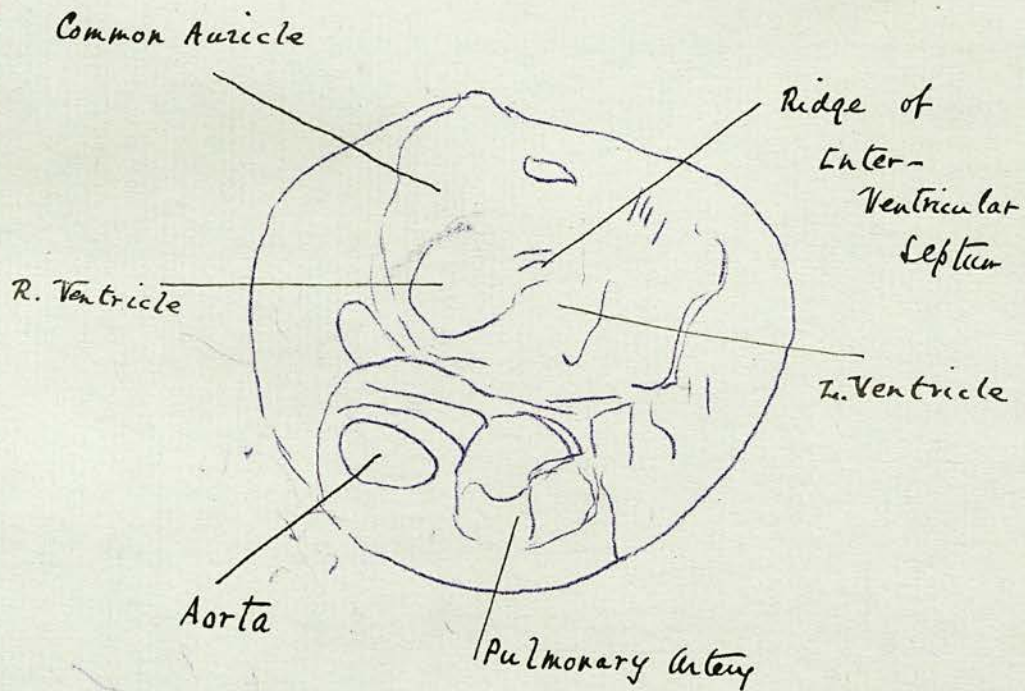
The respiratory system shows no abnormality. The respirations are 40 per minute. There are now no bronchitic sounds. There is some indrawing in the supra-clavicular fossae.

Digestion is now good. The liver is in its usual place and is not enlarged. The spleen is not palpable.

The urine is normal.

In brief the child is a clear example of dextro-cardia of congenital origin without transposition of the other viscera plus indications of cardiac disease or much more likely cardiac defect. This then is the record of the child's condition two months before death. There then seemed the promise of a year or two of life but the onset of whooping cough with bronchitis and convulsions brought about death at the age of $2 \frac{10}{12}$.

A partial post-mortem showed the liver in its normal position on the right side. The heart and pericardium were situated on the right side and stretched from the mid-sternal line to the right of the nipple line. The pericardium and the diaphragm were intact. There was no signs of adherent pericardium. The lungs were congested. There was a slight amount of /



Photograph of the Child's Heart showing relations of parts.

of fluid in the pericardial sac but no adhesions were present. The heart lay with its axis extending from above and at the sternum downwards and to the right to the right nipple and below it. The heart was enlarged. There was one common auricle without any trace of division into two. There was one auricular appendix - the right. The auricular wall was smooth. Both ventricles were dilated and hypertrophied. Both mitral and tricuspid valves were incompetent. There was a distinct vestibule leading from the common auricle to the ventricles. There seemed a deficiency at the upper end of the inter-ventricular septum but there was a distinct line of septum below marking off the two cavities quite sharply. Both the aortic and pulmonary valves were competent. The ^{front} part of the heart and the greater part to the right was made up of right ventricle.

The case-record just outlined presents for consideration two things - 1 the heart defects and 2 the heart displacement.

The Heart Defect. The condition described in this heart represents that type of heart defect which is known by the name cor triloculare biventriculare. Ziegler special pathological anatomy 1896 sections I - Viii mentions in detail the different names applied /

applied to hearts with septal defects. The three chambered heart may be formed with one auricle and two ventricles as in this case when it is called the cor triloculare biventriculare or else it may have one common ventricle and two auricles when it is designated the cor triloculare biatriatum. Both are rare and both are only found occasionally with such a complete absence of the septum as justifies the definite name. In short the tendency for all these septal defects is to vary so much in the actual defect as to prevent the occurrence of any pure type. They always tend to pass from one to the other so that one may have as the slightest degree at one end of the scale a patent foramen ovale up to the complete absence of the septum as in this instance.

The number of recorded cases of cor triloculare biventriculare is but limited. The chief examples that I have met with are -

1. Fussell. M.H. (Med. News, Nov. 3 1888) describes a case in a child aged 20 months who had a partial dislocation of the abdominal organs. There was no septum between the auricles and the ventricular septum was incomplete at the upper edge. There was only a common atrio-ventricular ostium for both ventricles /

cles.

2 Probyn - William J. of anat. & phys. vol XXViii part iii, 1894. p 305 - 308 under the title 'Unusual Malformation of the Heart' gives the report of a full time infant who was cyanosed from the first and who only lived one month. The apex of the heart was recognised during life as being to the right of the sternum. There were no murmurs. A patent foramen avale was suspected. The post-mortem showed a heart of normal size with an apex to the right of the mid-line. There was one auricle about the size of the two together. There was a small septum from the middle of the upper part of the posterior auricular wall, evidently an abortive attempt at septal formation. The auricle was separated from the ventricles by a vestibule common to the three cavities and at the bottom of it there was the ridge of the interventricular septum with openings on either side into the right and left ventricles. There were also various anomalies of vessels.

(3) Ewald (Berl. Klin. Woch. 1898. No. 47. p. 1044 - 5) describes the case of a man, aged 42, in whom he diagnosed a heart defect. The heart was dilated. Systolic & presystolic murmurs were present. There was distinct local cyanosis especially marked on the cheeks, ears /

ears and the fingers. A post-mortem showed a complete absence of the septum of the auricles and dilatation and hypertrophy of both ventricles. The patient's father and also his brother had had heart disease. Only three or four with such a defect had reached the 40 - 50 decade. Hanseemann in the subsequent discussion referred to a like heart in a man of 62.

(4) Langer (Prag. Med. Woch. 1899 No. 8) describes a very typical case in a patient with total transposition of the viscera.

(5) Thérémán (Études sur les Affections Congenitales du Cœur p. 154 and figs. 229 - 231. 1895) describes and figures a heart with complete absence of the auricular septum. The common auricle communicated with the two ventricles by a large opening separated in the midst by the septum of the ventricles.

The conditions found in all those cases can only find their explanation in a consideration of the manner in which the Septa are formed. Keith (Human Embryology and Morphology 2nd ed. 1904. p. 287) mentions how during the latter part of the first month and the opening of the second the auricular part of the heart becomes separated into right and left chambers by the formation and union of three Septa:- (1) the endocardial /

ial cushions; (2) the septum prim^{um}~~era~~; and (3) the septum secundum. In the case of the pure type of cor triloculare biventriculare there is a complete arrest in the development of these three septa and therein is found an explanation of the common auricle and also as has been shown lately of the common vestibule leading to the separate ventricles. A partial arrest can give a very varied anatomical type of defect of which the abortive septum in Probyn - William's case is an example. These embryological data thus give a full and adequate account of the conditions found in these hearts where there is this type of septal defect and no complicating valvular involvement. The slight gap at the upper end of the inter-ventricular septum is chiefly due to the absence of the endocardial cushions which go to form it. As regards the clinical picture in these cases it is difficult to lay down any definite rule as it is just hearts such as these that so often show valvular lesions which may of course give rise to signs that mask or at least complicate those produced by mere septal defect. Cyanosis seems to be the only sign that gives a suggestion of their presence. This cyanosis too seems in some cases only to develop in later life either after some bodily or mental strain or /

or else without any special cause. Such is the opinion of Ewald who in discussing his case lays special stress on the local character of the cyanosis.

The physiology of these hearts with such defects is somewhat obscure. It is presumed that there are two streams of blood running side by side and that there is very little mixture taking place except at the points of contact along the edges where the two columns of blood are in contact.

The Displacement of the Heart. The displacement of the heart to the right which this patient shows seems well worthy of detailed consideration as the number of cases of like nature is as yet but small and their nature is by no means understood. These right-sided displacements may occur from various causes and at various periods in the life history of the organism. They may be part of a general transposition of the organs of the body in which there is a complete reversal of the two sides of the body and of the parts of the individual organs. This constitutes situs viscerum inversus or as it is also called heterotaxia totalis. On the other hand there may be a transposition of the contents of one cavity either thorax or abdomen, when there results a partial situs viscerum inversus /

inversus. There are also not a few transition cases such as the one mentioned by Abernethy (3) in which there was along with an almost complete invasion of the thoracic organs a liver situated in the middle line. Still more curious and difficult to classify are cases such as that lately described by Royer & Wilson (Brit. J. of Children's Diseases vol. V. pp. 176 - 8) in which there are complete transposition of all the organs of the chest and abdomen except the heart which however was full of pathological faults.

Our knowledge regarding displacements of the heart has only grown very gradually. There is very little to be found in literature until about the 17th century but by the beginning of the 19th century there were isolated communications and even attempts to collect the recorded cases as was done by Bresschet (8) The early cases were naturally mostly post-mortem records and it is only about the middle of the century that clinical records began to appear. Schrötter (17) in 1870 reported a case and reviewed the subject. The study of the always attractive subject of general transposition of the viscera helped greatly in bringing to light the lesser transpositions and in Küchenmeister's work on this subject (25) there was much indirect and often critical consideration of these isolated /

isolated cases of inversion. Even before this Krieger (19) had been able to collect 16 cases of what he believed to be examples of this isolated dextrocardia. Withal there was a want of definition of the real meaning attached to the word dextrocardia. Löchte (50) in 1894 discussed the whole matter afresh in a frankly critical manner with a resulting diminution in what he considered true cases. At the same time he advanced new theories regarding its causation.

Since Löchte's paper the stream of communications has continued but it is questionable if any great progress has since been made. There have not been any fresh reviews of the subject excepting a short but comprehensive paper by Löwenthal in 1900. (70).

Congenital Dextrocardia may be defined as the name applied to a displacement of the heart to the right side in which as in general transposition of the viscera there results a mirror image of the heart corresponding to that on the left with at the same time a complete reversal of the vessels and the cavities of the heart. Now while this is the ideal definition which it is evident later observers have in their mind it is very questionable if any of the cases yet reported can be adduced as examples of this true congenital /

ital dextrocardia.

It is difficulties such as these that have led me to attempt to pass in a comprehensive review the varied kinds of cases that have been so designated in the hope that some kind of order may be evolved. It is first of all necessary to set aside all those cases in which the dextrocardia is part of a general transposition and confine one's attention entirely to displacements of the heart only - the isolated dextrocardia of the Germans, the pure dextrocardia of the French.

These isolated or pure dextrocardias may be found under varying circumstances and at different times in the life history of the organism. The classification that commonly holds good is into congenital and acquired but many facts go to show that a much more elaborate division must be made if one is to be able to appreciate the many difficulties that arise in the diagnosis and etiology of true dextrocardia. Acquired dextrocardia of post-natal life does not directly come into consideration in the paper except in relation to the diagnosis of the congenital form. Even, however, in connection with congenital dextrocardia it is very advisable to consider and classify the many cases that have been reported as such although it is very likely most of them will /

will have to be rejected as untrue examples of the condition. These congenital dextrocardias may be due entirely to changes in the heart itself or else they may have been influenced by changes in neighbouring structures. There is thus separated out two distinct groups of displacement:- one due to intrinsic changes and the other to extrinsic.

Group Extrinsic. It seems well to consider in the first place the group of congenital dextrocardias due to extrinsic or external causes. This type of dextrocardia corresponds largely to the acquired cases of post-natal life. Not a few of the cases reported as congenital dextrocardia come under this heading. They are certainly much more cases of dislocation of the heart and ought more properly to be called dextro-versio cordis as is now being done in some cases. They differ in their origin from the true dextrocardia of intrinsic origin and also show in their subsequent history many points that help to differentiate them.

These extrinsic cases can be classified according to their cause in the following way:-

- (1) Cases due to changes in the lung.
- (2) Cases due to diaphragmatic herniae.
- (3) Cases due to ante-natal inflammatory conditions /

ditions leading to displacement from adhesions as in the acquired cases of later life.

(1) Lung Change.

(a) The first cause of ~~M~~-sided displacement is complete absence of the right lung as in cases reported by Meckel (7) and by Maschka (14). In Maschka's case the whole of the right pleural cavity was filled up by the heart. Allied to this is the partial absence of the right lung as in a recent case reported by Berliner(92) in which there was a dextroversio cordis with non-formation of the upper and middle lobes of the right lung.

(b) A curious cystic transformation of the lung has in occasional cases given rise to dextrocardia. Krieger (19) in his important thesis on congenital dextrocardia describes such a case in which the subsequent post-mortem showed the presence of this condition in the right lung and thus excluded it from the category of true dextrocardia. These cystic lungs suggest clinically ^{cl}bronchiectatic cavities and show pathologically a cystic formation in the lungs with absence of pigment. Carpenter (82) has recently published with illustration a case in which a large cystic left lung appeared to have led to a dislocation of the heart /

heart to the right. Kreisch (48) in a case with dextrocardia and lung conditions suggesting this condition attempts to argue its presence.

(c) There may be malformation of the lobes of a lung leading to enlargement of a lung and displacement of the heart. This occurred in a case reported by Krönig (66) in which the left lung was so affected. The dextrocardia was diagnosed during life. There were no heart anomalies beyond this.

(d) There may be atelectasis alone or else it may be along with misplacement. Such a case is that of Perregaux (51) in which an eight month child with dyspnoeic attacks was found to have its heart beating in the 6th rt. intercostal space in the nipple line.

There was no murmurs heard on the heart. ^{Breathing} ~~Hernia~~ was almost completely absent over the right side. A tumour of the mediastinum was suspected. A subsequent post-mortem by Morestin (52) showed the heart in the right side of the chest and the right lung flattened and misplaced.

(2) Diaphragmatic ~~Hernia~~. This has long been recognised as a cause of lateral displacement of the heart to the right side when the ~~hernia~~ occurs as it usually does on the left side. Stokes in 1854 in his Book /

Book on Diseases of the Heart and Aorta mentioned the condition as being most common in new-born infants who only lived a short time but also cited examples to show its presence was not incompatible with a considerable duration of life. Thus in a case of Weyland quoted by Bouillaud the child which lived for seven years although liable to continual vomitings from the first period of its existence showed post-mortem the left side of the chest as high as the 2nd rib filled with the convolutions of the intestines and the lung only one-sixth of its ordinary volume. The heart was situated in the middle line. Two others are cited in one of which ^{from} ~~has~~ Gruveithier (17th Livraison of this Pathological Anatomy) the patient, a woman of 65 had the heart completely displaced to the right. Like cases have been reported by Alexander and Bailey (16). (10), by Guftmann (31). Lately I had the opportunity of seeing a new born infant which showed a distinct dextrocardia, the whole heart being to the right of the middle line. The heart sounds were normal. The child breathed quickly and seemed dyspnoeic. There was no abnormal sounds heard over the right lungs. On the left side there was dulness and the heart sound was absent. But for the age of the child one would have /

have suspected pleural effusion on the left side. An X-Ray photograph showed a dulness on the left side of the chest. In view of the age of the child the case appeared due to some mediastinal tumour. The child only lived a few days and at the post-mortem the heart was found on the right side while the whole of the left side of the chest was taken up by liver and intestinal coils which had entered by a hernia in the left side of the diaphragm.

Ballantyne (Antenatal Pathology & Hygiene, 1904 pp. 477 - 487) in a review of 100 recently reported cases of congenital diaphragmatic herniae states (p. 483) that in 9 the heart was pulsating and displaced to the right.

It is thus abundantly evident that the possibility of such a cause of congenital dextrocardia must be kept in mind.

(3) Ante-natal inflammatory condition.

There seems to be no doubt regarding the existence of these and of course if these inflammatory processes affect the right pleura and lead to adhesion between it and the pericardium the possibility of displacement such as occurs in the acquired dextrocardia of later life is evident. In such cases congenital dextrocardia /

cardia may be diagnosed when they really are cases of dextroversio due to inflammatory processes occurring during foetal life. Unless one had a very clear record of an infant's health especially during its early years it would be difficult in later years to estimate and name correctly a dextrocardia discovered in later years and even a post-mortem examination might only confirm the presence of an acquired displacement without helping to settle whether it was ante-natal or post-natal in origin. Thus Cooper (11) in 1836 described the case of a child with distinct dextrocardia, the sequel of a right-sided pleurisy probably of foetal origin or else occurring in the first days of life. The child was ~~cyanosed~~ cyanosed but lived 10 months. Küchenmeister (Op. cit. p. 244) mentions the more convincing case of a lithopaedion with the heart to the right and with adhesions between the right lung and the pericardium.

This concludes a survey of the chief factors at work in the production of cases of dextrocardia due to extrinsic causes. Paltanuf (76) especially pleads for this type of dextrocardia being designated dextroversio. With post-mortem findings their definite nomenclature is easy but without that many of them pass as cases of true congenital dextrocardia.

Group II. Intrinsic.

The second great group of cases of dextrocardia consists of displacements of the heart without any evidence of such disease defect or malformation in neighbouring structures as might cause this lateral movement. This group includes all the cases which everyone recognises as true congenital dextrocardia. These cases may have been discerned only post-mortem, or else they may have been diagnosed during life and confirmed after death or lastly their supposed nature may only rest on a clinical basis. There is thus set out for consideration these three kinds of cases.

(a) Cases found only at post-mortem.

This group of cases is formed mostly by the earliest recorded cases before physical diagnosis had taken its place in the examination of the sick. The list of such cases is not a large one if the exclusion of cases of total transposition of all the viscera or even of cases with partial transposition of abdominal viscera be rigidly adhered to. There would thus be set aside the case of Mollenbrok (1) where the stomach lay to the right as well as the oft quoted case of Sampson (2) where the liver lay in the left hypochondrium and the spleen in the right as well as the transition case of Abernethy's (3) where the liver lay in the /

the middle line and even some others where the absence or meagreness of the reports regarding the position of the abdominal organs debars them from inclusion here. The earlier recorded cases centre around the names of Otto and Breschet but at the present time rigid criticism of these by many observers has done much to limit their numbers. In one case of Otto's (5) the heart merely lay a little to right. The patient had phthisis but apparently the conditions were not such as to produce any changes on the position of the heart. 1. Otto's other case (4) concerned a child with a right-sided position of the heart, The vessels were as usual as regards their position. There was a defect in the septum ventriculorum.

2. Berschei (18) reports the case of a boy who died at 1/12th. The stomach and liver were normal in position. The right sided heart showed defect in the septum of the auricles, absence of the septum of the ventricle, and the aorta came from the common ventricle.

3. Robinson (A.R.) (22) reports a case of transposition cordis with cor biloculare.

4. Kundrat (27) mentions the post-mortem report on a boy of 5/12ths with a right-sided position of the heart and with transposition of the large vessels. The aorta came from the anterior right cavity and the pulmonary /

monary from the posterior-left. The pulmonary artery was narrowed. The Ductus Botelli was open.

5. Löchte (50) describes in detail the pathological condition found in a man who died at the age of 46 from arterio-sclerosis and chronic myocarditis. The right-sided heart showed a transposition of the ventricles and there was also a corrected transposition of the large vessels so that in spite of the transposition of the cavities the large vessels came from the cavities they belonged to.

6. Baumgarth (79) notes a case of cor biloculare in which there was also a dextrocardia.

A detailed consideration of these cases will come more naturally along with the next group where there was in addition to the pathological report the recognition of the condition during life.

(b) Cases with both clinical and pathological report. This group is not by any means large and of course the reported cases come at a later date when the discovery of the methods of physical examination allowed and led to a more careful and also more accurate estimate of the contents of the chest.

Lowenthal (70) who in 1900 separated out this definite group considered the case he reported as only the fifth published one but as one of his cited cases is /

is that of Krönig (66) where the dextrocardia was associated with abnormal breadth of the left lung it seems more to belong to the previous division of extrinsic dextrocardia. Since Lowenthal's papers there have been very few cases that fall under the heading. Cipriani (36) reports a case of a man whose dextrocardia was recognised during life but whose heart with defects of both septa was adherent to the pericardium and to the right lung. The condition was not recognised at birth and though there is a report of an illness in early life it still remains a moot question whether the dextrocardia was acquired or whether a person with congenital dextrocardia had added to it a later pericarditis and pleurisy. A somewhat similar line of argument is applied by another observer Paltauf (76) who found a pericarditis at the post-mortem of a case which had been described years before by Bamberger (30) as a congenital dextrocardia. The only other observation on record is an interesting case of Pal's (87) which gave in 1907 a pathological confirmation of a case shown and demonstrated in 1888 by Gruss (29) at the same time as the case of Bamberger's. My own personal contribution seems a case fitting in with the requirements of this section.

The /

The list of such cases is the following:-

1. Pope (24) A man of 41 who died of pneumonia, had during life his apex beat 2" below the right nipple and 2" to the right of the sternum. There was a mitral regurgitant murmur. There was no history of pleurisy, empyema or pericarditis. At the p.m. the heart was found nearly entirely to the right of the middle line. There was mitral endocarditis. There was no pleural fluid and there were no pleuritic adhesions. The vessels were normal.
2. Grunmach (35) this case concerns a boy of 15 who had been dyspnoeic and cyanosed since his second year. The post-mortem showed dextrocardia without transposition of the vessels. The foramen ovale persisted. There was pulmonary stenosis. There was a foramen in the septum ventriculorum.
3. Graan-boom (37) reported a dextrocardia with the apex beat in the rt. anterior axillary line at the 6th rib. The detailed post-mortem by Reddinguis (32) showed a complete transposition of the vessels.
4. Lowenthal (70) The A.B. was in the 5th rt. space. The p.m. showed an absence of the pulmonary artery with the ductus Botalli as arterial vessel. There was a gap in the upper part of the septum of the ventricles.
5. /

5. Pal (87) This patient was reported as a dextro-cardia ~~by~~ Gruss (29) in 1888 and had again come under observation about the time of her death in 1907. The heart was noticed to be beating on the right side when the child was 3 days old. The A.B. was under the edge of the 4th rib a little outside of the nipple. There was a systolic murmur. The p.m. confirmed the dextrocardia. There was also aortic stenosis. The large vessels ~~were~~ unchanged.

6. This is the personal observation detailed ^{above} ~~along~~ with a dextrocardia observed from birth and a p.m. showing a cor triloculare biventriculare and no change in the large vessels.

III. Clinical Cases only.

The largest and last group is made up by those with only a clinical report and as it is important to see the kind of case which is put forward as dextrocardia an attempt will just be made to put very briefly the main and apparently salient points of each individual ~~one~~ in tabular form:-

Author	Sex	Age	Heart	Lungs	Other Remarks.
Mosler (15)	M	20	Mirror type	normal	
Schrotter (17)	M	32	A.B. 5thrt. axillary space.	Rt. Lung impaired above. absolute dulness below	Congenital dextrocard- ic + added traction.
Mosler (18)	F	36	Mirror type	normal	
Leichten- stern (19)	M	21	Patent for- amen ovale pulm. sten- osis.		Heart disease from birth
Henoch (19)	F	11	Pulsation 3rd rt. space.		Rt. index only two phalanges. Absent right thenar.
Bramwell (20)	M	39	Pulsation 1" outside rt. nipple line.		
Schrötter (26)	M	22	A.B. under sternum. Heart also to rt. of sternum.	Left pleural exudate.	
Sussmann (28)		15	Free movements of heart and lung.		Pleurisy, early childhood.
Grünfeld (33)	F.	44	A.B. 5th 2 c.m. inside A. 1.		Noticed at 14.
Niesel /					

Niesel (34)	M	20		Rt. sided pleurisy	Author uncert- ain.
Becker (38)	M	25	Mirror type 4th.		No previous illness.
Hawkins (40)	M	12	A.B. bet. 5th & 6th 1" out- side N.L.		Pulm. stenosis and regurgita- tion.
Ewart & Bennett (41)				bronchiec- tasis.	
Bard (42)	F	15	Copy of opposite side		Mitral sten- osis.
Heimann (43)	M	21			Left half body and left eye smaller. Left temple hollow. Chest also mis-shapen.
Carmich- ael (45)	M	7	Systolic occasionally		Bronchitis and Emphysema previously.
Gerrard (54)	F	26	A.B. 4½ to right	Never had pleurisy	Mitral sten- osis and re- gurgitation.
M'Lennan (55)	M	15		Weak breathing over right lung.	
Berks (56)	M	3½	Pulsation 3rd, 4th & 5th.		Cyanosis.
Bramwell (57)		5			Pulm. stenosis
Vehsemeyer/					

Vehsemeyer (59)	M 16	A.B. 4th right in- side n.l.	Lung changes on left side	X-ray.
Sobierajczyk				
Wardrop- Griffatt (61)	F 7	A.B. 3rd right		Pulm. sten- osis + Septal de- fects.
Petit et Ravaut (64)		Heart normal		Recognised at 13
Middleton (65)		Heart normal		Absence left radius and thumb. Curvature of thorax. Left chest small.
Leo (68)	M 8	Heart normal	No lung disease	
Chapman (69)	M 40	A.B. 4th n.l.		No history of pleural pericard- ial or lung disease.
Crispino (72)	55		No chest affection.	Recognised since 15th year.
Bonheim (73)	M 7	Pulm. sten- osis + other de- fects		
Schmilinsky (74)	F 9	Ductus Botalli persistent		A twin. Noted from birth.

Monks /

Monks (75)	M	28	Heart normal	No previous illness.
Wagner (78)	M	23		Asymmetry of face on right side.
Weinberger (77)	M	19	Aortic stenosis.	
Flatau (80)	F	37	Mirror type	No previous illness.
Neumann (84)	M	20	A.B. 5th	phthisis rt. apex.
Hawthorne (86)	M	11	Systolic at A.B.	
Tate (88)				Phthisis.
Carpenter (90)				Few moist sounds rt. base.

These three groups of congenital dextrocardia are naturally of different value and cannot lead to the same aim of certainty in diagnosis. The first two where the pathological lesion is known must form the basis of any consideration regarding either the diagnosis of the condition or the discussion as to its origin.

In all of them the age is very varied and is really of no importance as it obviously depends on the associated lesions of heart lungs or other organs and not on the displacement as such. It also seems idle to analyse the incidence of sex here. The cases founded only on clinical observation provoke criticism in many ways but instead of dealing with them individually it seems better to make use of the knowledge gained ^{from} ~~for~~ the consideration of all the possible causes of congenital dextrocardia as well as from the records of pathologically confirmed instances in arriving at a means of correct diagnosis. Incidentally this will allow of a general criticism of the claims put forward for these clinical cases.

The diagnosis of congenital dextrocardia depends on (1) the recognition of the dextrocardia and (2) the proof of its being congenital. Dextrocardia is recognised by the absence by all the usual clinical methods /

methods of a heart on the left side and its presence on the right side. This, needless to say, implies in straightforward cases that an apex beat or presumably such is beating on the right side and can be confirmed by palpation that percussion supports the underlying presence of a heart and that auscultation gives heart sounds of such strength and tone as such a change in position would lead one to expect. All this is generally described in typical mirror cases but of course it may be modified if the heart is merely a little to the right of the middle. The cases described above respond quite readily to these methods of examination which have of course been applied to their solution. At the same time these methods have their limitation as cases presumably dextrocardia have pathologically been found to be merely enlargements of the heart to the right. Further these methods without a complete anamnesis cannot exclude a lateral displacement of considerable degree. Bard (42) has tried to show that ordinary lateral displacement gives a pulsation at a higher level than would a mirror type of heart and that the base is the most moveable part of the part.

Schrötter and others after him have laid stress on /

on the accentuation of the 2nd sound at the base on the right side in true dextrocardia but unless as a supplementary sign it seems unworthy of such stress being put upon it.

The other means of determining the presence of a dextrocardia is by the newer X-ray method. It was first used in this connection by Wehsemeyer (59) and at later dates by Senator (67) Petit et Ravant, Leo, Crispino, Weinberger, Neumann. Pal, Tate, Grunmach and the personal observation herewith. While it confirms the dextrocardia its most hopeful use is in determining the axis of the heart. In real congenital dextrocardia the axis should run from above and left downwards and to the right and not as in mere lateral displacement with so often merely an axis parallel with its original or only slightly less sloped to the right. Some also think they can determine the course of the vessels.

As a necessary sequel to the recognition of the dextrocardia there is the need to exclude any general inversion of the other organs. A lack of this as has already been mentioned invalidates many of the older cases. In the case of Carmichael reported above there was some question as to the position of the liver and the /

the spleen and this, apart from other things, makes its inclusion here doubtful. The second point is to determine the congenital nature of the displacement. As will be noted the right-sided position of the heart from birth was only noted in a very few instances. Where it was so observed the next point obviously is to settle where it is of intrinsic or extrinsic origin. This in itself is no easy matter and many cases reported as true dextrocardia have at a later date been shown to be due to the extrinsic causes detailed previously. If there be no note as to the condition from birth then a proof of its congenital nature has to rest on a personal history free from such chest conditions as might cause displacement along with a physical examination that supports this contention. The especial points requiring establishment are the free movements of the heart and the lungs, equal respiratory murmur on the two sides and absence of any change in the shape or size of the chest that might be suggestive of past disease. An X-ray examination is also useful here in showing a healthy lung condition and in defining the level and the movements of the diaphragm and thus helping to exclude past pleurisy or pulmonary retraction. This method of arriving at a diagnosis /

diagnosis is the one that justifies the name 'congenital dextrocardia' being applied to cases such as the two cited by Mosler, the first case of Bramwell's, those of Becker, Chapman and others mentioned above. It is however when congenital dextrocardia is diagnosed in cases with the simultaneous presence of pleural or pulmonary disease as is so often done in the above list that one is entitled to accept it only with a very becoming suspicion. Without a history of the presence of the heart displacement being anterior to the chest condition post-mortem records have in most cases shown that the diagnosis of its congenital nature is wrong. Acquired dextrocardia is moderately common and it occurs just subsequent to pleural or pulmonary affections such as are detailed above while congenital dextrocardia is one of the rarities of medicine and can only be diagnosed where every possible cause of the acquired forms can be excluded. As a case in point is that described by Berwald (44) as a congenital dextrocardia. It concerned a man of 17 with no illness of any consequence previously. The a.b. was at the right side just inside the right nipple line. The patient died from phthisis but there was no adhesion between the pericardium and the right lung. The apex beat was directed to /

to the left. There was some adhesion between the sternum and the pericardium. The cases must thus be considered an acquired one.

Two interesting facts emerge for consideration before the question of the etiology is approached. The first is the great frequency both in the post-mortem cases and even in the clinical cases with which defect or disease of heart is present. There is indeed nothing so striking in the whole series of cases as the close relation which exists between the displacement and the disease or defect of the heart. The nature of the cardiac lesions varied considerably but far the greater number were septal defects in either cavity or in both. In some cases there was absence of vessels. In ^{heart} ~~that~~ arrest of development rather than disease is the prevailing thing. Hochsinger (Pfaundler & Schlossmann, The Diseases of Children, vol. iii. p. 486) bears this out when he says "The congenital changes in the position of the heart whether accompanied by situs inversus or not are almost always associated with intra-cardial arrest of development." Indeed if one were to argue from the cases of congenital dextrocardia with pathological control one would be inclined to lay some stress on the presence of signs of congenital /

ital heart disease as, other things being favourable, an argument for the congenital nature of the dextro-cardia.

The other thing of interest is the presence of defects elsewhere as in the case of Henoch with a right index with only two digits and an absent thenar^r eminence, or the asymmetry noted in the cases of Heilmann and Wagner, or lastly the various defects noted in Middleton's case.

The last point for consideration is the ultimate causation of these causes. Many have offered explanations and have usually applied to these cases of partial inversion the same theories that are brought forward as solutions of general inversion. It is, however, very doubtful if they have any bearing in this relation for strange though it may seem to say it there has not yet been described any single case of congenital dextro-cardia which will fit into the definition put forth in the beginning. None of these congenital dextrocardias are true inversions and as has been seen almost all are complicated with heart defects. Even where the cavities have been reversed the vessels are corrected so that Schrötter (26) and Paltauf (76) are justified in their view as to the absence as yet of any true case of /

of inversion of heart and blood vessels. A. K. Stone (81) in a short review of the subject also comes to the conclusion that almost all the known cases of dextrocardia have been accompanied by extensive malformation of the heart and that no true case is on record.

The explanation of these cases such as they are has been attempted along different lines. In the slight cases of displacement it has been assumed that the presence of heart defect has led to an enlargement of the heart at an early date in foetal life when the heart is still mesial in position. Lochte (50) considers these heart anomalies never occur without some abnormal arrangement of the vessels or organs of the abdomen. The inverted position of an organ such as the heart is due to some local developmental anomaly and the presence of so many pathological faults in these inverted hearts goes to support this. It also indicates its occurrence at an early stage of development i.e. before the second month when the septa close. The present case with its three generations with heart disease would thus seem in its displacement and defect to indicate a weakening in the germinal area concerned in the development of the heart.

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of

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